



# Family Health *Dataline*

## IN THIS ISSUE:

- On average, 18% of children born each year in Alaska are reported to the Alaska Birth Defects Registry with at least one reportable birth defect (birth years 1996 – 2000). About 5% of children in each annual birth cohort are reported with at least one *major* congenital anomaly.
- Timeliness of reporting and early case ascertainment has improved since inception of the Registry in 1996. Approximately 61% of birth defect reports submitted to the ABDR in 2001 were received within 150 days of diagnosis or treatment.
- Reporting lag time and diagnosis of birth defects detected in older children, along with a declining birth rate in Alaska, accounts for an observable decrease in the number of children reported for each birth cohort after 1998. Estimation of birth defects prevalence and epidemiological analysis may be delayed if there is incomplete case ascertainment for a given birth year.
- Initial verification studies of surveillance data showed that the predictive values of the ABDR were high for four of seven anomalies reviewed and that predictive value varied by reporting source.

## Surveillance Update: The Alaska Birth Defects Registry 1996-2002

### *Background*

Alaska Statute established the Alaska Birth Defects Registry (ABDR or Registry) in January 1996. In the January 1999 issue of the *Family Health Dataline*, we published preliminary information on the prevalence of major congenital anomalies among Alaskan children born in 1996 (1). Since that initial analysis was conducted, ABDR staff have enhanced surveillance efforts to improve birth defects reporting and to identify and address data quality issues.

An important component of any public health surveillance program is to periodically monitor and evaluate the system to ensure that it is efficient and effective (2). High quality registry data is a prerequisite to reporting information on birth defects prevalence and for using ABDR data for epidemiological studies. In preparation for publishing our next report on the prevalence of major birth defects in Alaska, we evaluated the ABDR for completeness, timeliness, and quality of data. In this issue of the *Family Health Dataline*, we describe our findings and make recommendations for improved birth defects surveillance for Alaska.

### *Methods*

Previous issues of the *Family Health Dataline* have described in detail the surveillance methodology and reporting guidelines for the ABDR (1,3). Reportable birth defects include International Classification of Diseases, 9th revision (ICD-9) codes 740-760, certain metabolic disorders, infantile cerebral palsy, muscular dystrophy, various anemias, congenital hypothyroidism, and neurofibromatosis. Physicians, hospitals, and other health care facilities and providers are required to report children from birth to one year of age who have any of the reportable birth

defects. Children treated or diagnosed with conditions defined by ICD-9 codes 760–760.9, including those related to maternal alcohol use and Fetal Alcohol Syndrome (760.71), must be reported until the 6<sup>th</sup> birthday (4). The ABDR relies on multiple-source reporting from health care providers statewide, and the ABDR database is structured to allow for the linking of multiple ICD-9 codes and reporting sources for each child. To ensure that an unduplicated count of children is maintained by the ABDR, and to obtain additional epidemiological information, birth defect reports are routinely linked to vital records.

In 1999 through 2000, ABDR staff implemented several enhancements to birth defects surveillance. First, we developed an improved data management system in Microsoft Access. This improved the efficiency of data storage and retrieval and allows for more comprehensive evaluations of reporting sources. We also programmed systems for electronic importation of reportable birth defects from large data files. This reduced manual data entry time and has allowed sources to report in electronic formats.

To improve timely compliance with birth defects reporting regulations and to ensure full participation, ABDR staff implemented various methods of providing information to health care providers. We traveled throughout the State to inform providers and medical records personnel about the ABDR reporting requirements and reporting methods.

We developed data entry protocols and a procedure manual (with written quality assurance guidelines). In 2001 and 2002, we also increased staff capacity to allow for more timely data entry, data verification, epidemiological analysis, and dissemination of findings. By 2002, all 24 major hospitals in the State were reporting regularly to the ABDR. During 2002, we began conducting medical chart reviews at selected facilities to validate reports submitted to the Registry and to assess the predictive value of those reports.

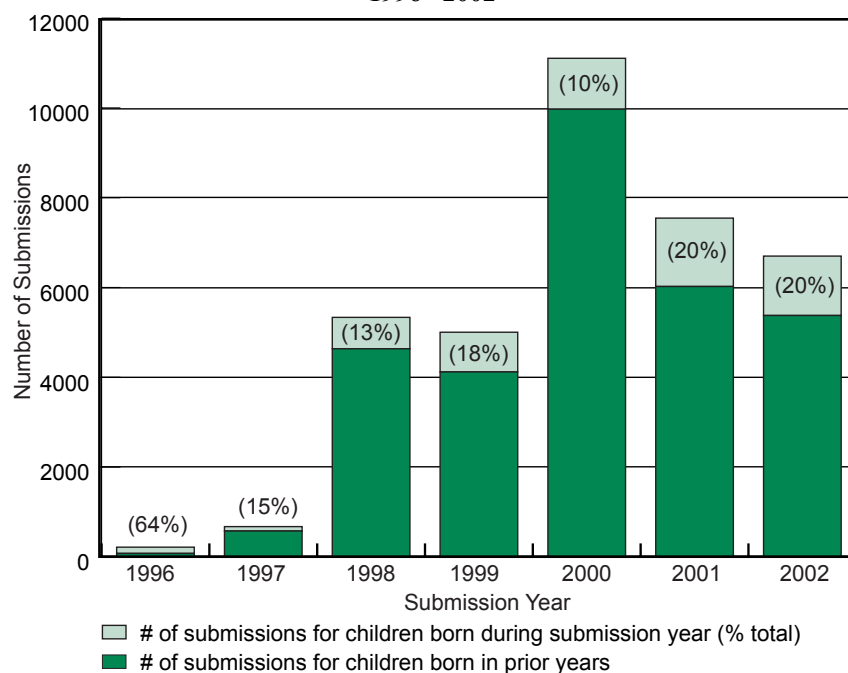
For this evaluation of the ABDR, records with unknown report dates were excluded from analysis.

## Results and Discussion

### *What improvements in reporting have occurred since the implementation of ABDR?*

Reporting to the ABDR has shown substantial improvement since the Registry was established in 1996, with improvements particularly evident in year 2000. A total of 11,106 reports were received in 2000, compared to just 674 reports in 1997, 5335 reports in 1998, and 5004 in 1999 (Figure 1). The increase in reporting to the ABDR during 2000 may be attributable to provider education efforts that began in 1999. After 2000, the number of reports submitted to the ABDR declined, possibly because many of the reports submitted in 2000 were “catch up” reports for children born in prior years. (It is

**Figure 1. Number of Birth Defects Reports Received by the Alaska Birth Defects Registry, by Submission Year, 1996 - 2002**

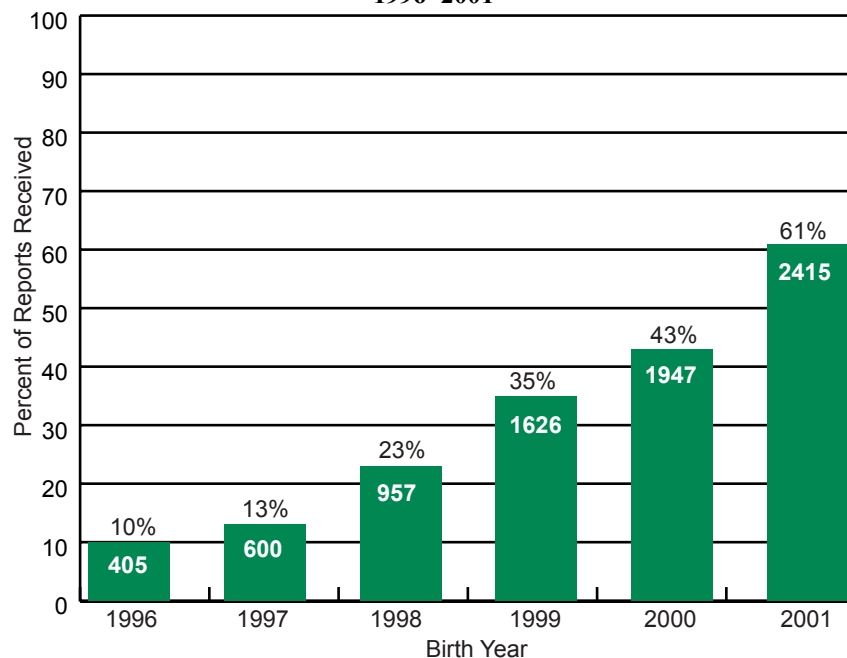


important to note that a child may be reported to the ABDR by one or more reporting sources; thus, the number of reports received by the Registry reflects the completeness of reporting as well as the number of children reported with congenital anomalies).

Health care providers are asked to submit quarterly all reportable conditions that are diagnosed or treated during the previous three months. We evaluated the lag time between diagnosing or treating a child with a reportable birth defect and reporting it to the ABDR. Using 150 days as the criteria for timely reporting, we found that the proportion of reports received by the ABDR within 150 days of the reported diagnosis or treatment date increased for each successive birth cohort (Figure 2). For children born in 2001, approximately 61% of birth defects reports were received in a timely manner, compared to just 10% for 1996.

While timeliness of submitting reports to the ABDR is improving, these data show that at least 40% of reports for children born in 2001 were not received within 150 days of diagnosis or treatment. Late reporting of diagnosed birth defects can result in several years of delay before an accurate prevalence estimate can be calculated for a given birth cohort. Full participation in birth defects reporting by all health care providers and timely submission of reports is critical for effectively tracking the prevalence of birth defects, for using surveillance data to evaluate the effectiveness of prevention efforts, and for conducting cluster investigations.

**Figure 2. Proportion of Total Reports Received by the Alaska Birth Defects Registry Within 150 Days of Diagnosis, by Birth Year, 1996-2001**



### *How consistent is ABDR data?*

For birth years 1996 through 1998 the number of children reported to the ABDR is relatively consistent (Range: 1903 to 1946). Epidemiological analysis of these birth years has begun and prevalence estimates for major congenital anomalies will be presented in upcoming issues of the *Family Health Dataline* (major anomalies are those that are defined and monitored by the National Birth Defects Prevention Network and reported annually in the journal of *Teratology* (5)).

The number of children reported to the Registry has declined for each successive birth year after 1998 (Figure 3). This observation is explained by the combined effects of reporting lag time, later diagnosis of birth defects detected in older children and a declining birth rate in Alaska. Whether or not a decline in specific birth defects over time has also contributed to the decrease in the overall number of reported birth defects awaits further evaluation.

There is considerable consistency for all birth years in the proportion of children reported with at least one major congenital anomaly. For each annual birth cohort from 1996 to 2001 the proportion of children reported with at least one major anomaly ranged from 27% in 1996 to about 31% for children born in 2001 (or an average of 28% for all birth years combined). Based on data received for birth years 1996 – 2000, our most complete years of data, an

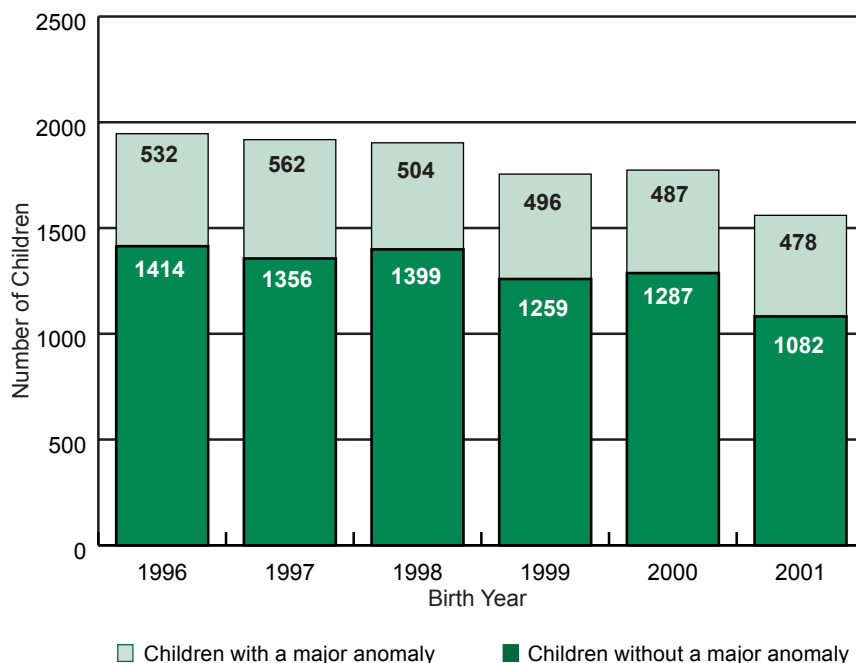
average of 516 children, or 5% of the approximately 10,000 children born every year in Alaska, are reported to the ABDR with at least one major congenital anomaly.

### *How well do reports made to ABDR reflect the actual occurrence of specific birth defects?*

The ABDR was established as a passive surveillance system. Essentially, this means that we rely on health care providers to report the diagnosis or treatment of a reportable birth defect to the Registry. ABDR staff do not actively search medical records for specific diagnoses, nor do they routinely confirm reported diagnoses (with the exception of ICD-9 Code 760.71; “fetus or newborn affected by alcohol via placenta or breast milk” and select birth defects).

We compared preliminary prevalence estimates for major congenital anomalies among children born in 1996 – 1998 to rates for states with well-established birth defects surveillance programs. Initial results showed that Alaska’s rates for some major congenital anomalies appear to be significantly higher than rates published by other states. Variability in surveillance methodology may explain some of the observed differences, however other surveillance effects such as over- and under-reporting, mis-coding of reported diagnoses, and reporting errors could also affect the validity of ABDR data. During 2002, we began conducting

**Figure 3. Number of Children Reported to the Alaska Birth Defects Registry with and without Major Anomalies\*, by Birth Year, 1996 - 2001**



\*For this analysis, "major anomalies" are those defined and monitored by the National Birth Defects Prevention Network.

medical chart reviews to validate reports submitted to ABDR and to estimate the predictive value of the surveillance system for selected birth defects.

We verified case reports for seven reportable conditions at two of Alaska’s largest hospitals. Birth defects chosen for chart review were those we suspected of being over-reported to the ABDR, or those with significantly higher rates in comparison to other states. The positive predictive value (proportion of reported cases that were true positives) for four of the seven sentinel conditions was over 90% (Table 1). Predictive values varied greatly by specific birth defect. Pulmonary valve stenosis/atresia had a low positive predictive value, indicating that most reports did not reflect the actual occurrence of disease. Both hospitals had information in the medical record that ruled out this diagnosis. For sickle cell anemia/sickle cell trait, cases were positively confirmed in only 14% of reported children, with a majority of the reviews resulting in an inconclusive finding, and thus, warranting further review.

Conducting verification studies of reported cases is important to understanding the validity of the Registry data. Information gained from chart review can be used to evaluate surveillance methods, identify needs for provider education, and provide information for statistical adjustment of rates. Our evaluation has identified limitations to the interpretation of ABDR data and has identified new directions for improving data quality.

**Table 1. Positive Predictive Value of Select Birth Defects Reported to the Alaska Birth Defects Registry Using Medical Chart Review as Gold Standard\*; Birth Years 1996-98**

Birth Defect	Confirmed	Ruled Out	Inconclusive	Total Reported	Positive Predictive Value
Pyloric Stenosis	47	1	2	50	94%
Gastroschisis	12	0	1	13	92%
Omphalocele	2	0	0	2	100%
Pulmonary Valve Stenosis/Atr- esia	12	30	2	44	27%
Neural Tube Defects	6	0	1	7	86%
Hirschsprungs Disease	12	0	1	13	92%
Sickle Cell Anemia/Trait	3	0	18	21	14%
Total:	94	31	25	150	63%

\*Case verifications were performed for these seven reportable conditions at two of Alaska's largest hospitals.

## *Recommendations*

- Continue provider education to encourage timely and more complete reporting.
- Periodically monitor timeliness and completeness of provider reporting and establish a system to give feedback to health care providers showing the timeliness and completeness of reporting.
- Create a standard procedure and reporting manual for health care providers and health information staff to provide detailed guidelines on birth defects reporting.
- Complete analysis of major birth defects prevalence in Alaska for children born in 1996-2000.
- Begin annual reporting of birth defects prevalence.
- Identify an appropriate source of data for comparison with Alaska rates.
- Continue and expand medical chart reviews to ascertain the validity of birth defects reporting.

*Submitted by Lisa Allen and Janine Schoellhorn*

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